Hemodynamic Profile After the Norwood Procedure With Right Ventricle to Pulmonary Artery Conduit

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Background—The balance of systemic, pulmonary, and coronary blood flow after the Norwood operation for hypoplastic left heart syndrome (HLHS) is critical to early survival. We hypothesized that a right ventricle to pulmonary artery conduit (instead of a systemic to pulmonary artery shunt) would result in hemodynamic changes consistent with a more stable balance of systemic, pulmonary, and coronary perfusion.

Methods and Results—Hemodynamic data were obtained during cardiac catheterization before the hemi-Fontan procedure from 24 patients with HLHS: the first 10 had a Norwood operation with a systemic to pulmonary artery shunt, and the latter 14 had the Norwood operation with a right ventricle to pulmonary artery conduit. Significant differences were present, with the right ventricle to pulmonary artery conduit group having a higher aortic diastolic pressure (55 versus 42 mm Hg), a narrowed systemic pulse pressure (43 versus 64 mm Hg), a lower Qp:Qs (0.92 versus 1.42), a higher coronary perfusion pressure (46 versus 32 mm Hg), and a higher ratio of pulmonary artery diameter to descending aorta diameter (1.51 versus 1.37).

Conclusions—We conclude that, in HLHS after the Norwood operation, the right ventricle to pulmonary artery conduit modification produces hemodynamic changes consistent with improved coronary perfusion and a more favorable distribution of systemic, pulmonary, and coronary blood flow. (Circulation. 2003;108:782-784.)

Key Words: surgery • hemodynamics • heart defects, congenital

Survival after the Norwood operation for hypoplastic left heart syndrome (HLHS) requires an appropriate balance of the systemic, pulmonary, and coronary circulations. Significant early and late mortality has been associated with a maldistribution of blood flow from the single right ventricle. Coronary perfusion may be at particular risk because of the diastolic runoff associated with a systemic to pulmonary artery (BT) shunt. A review of 122 postmortem cases after the Norwood procedure at the Children’s Hospital in Boston indicated that the most important causes of death were impairment of coronary perfusion (27%), excessive pulmonary blood flow, allow for appropriate pulmonary artery growth, and not interfere with the systolic or diastolic function of the right ventricle. To assess these hypotheses, cardiovascular hemodynamics at catheterization before the hemi-Fontan procedure were compared in 2 groups of patients with HLHS.

Methods

Two groups of patients with HLHS who had undergone a Norwood operation as newborns underwent hemodynamic evaluation by cardiac catheterization at ~5 months of age, before a hemi-Fontan procedure. Patients were accumulated over a 2-year interval (2000–2001) in 2 centers using the same protocol for perioperative management. During this time, no modification other than the source of pulmonary blood flow was introduced. Ten patients underwent a Norwood operation (transection of the main pulmonary artery and allograft with homograft and atrial septectomy) with a BT shunt (2000). Fourteen patients underwent a similar Norwood operation except for placement of an RV-PA conduit as the source of pulmonary blood flow. This conduit was a 5-mm polytetrafluoroethylene tube sutured from the right ventricular free wall to the distal main pulmonary artery stump in all but 1 patient (2001) (see Figure). All patients in the BT shunt group and nine in the RV-PA group were operated on at the duPont Hospital for Children. The remaining 5 RV-PA patients underwent surgery and catheterization at the Polish-American Children’s Hospital in Krakow, Poland. Cardiac catheterization was performed using standard techniques under moderate sedation or under general anesthesia if interventional procedures were anticipated. For both groups, pulmonary artery pressure was measured using 3F pigtail catheters. Calculations for
Qp:Qs were performed using the Fick principle and an assumed oxygen consumption of 180 mL/min · m². To compare pulmonary artery growth between groups, a ratio was obtained by dividing the sum of the diameters of the right and left pulmonary arteries by the diameter of the descending aorta at the level of the diaphragm in systole. Coronary perfusion pressure was obtained by subtracting the mean right atrial pressure from the aortic diastolic pressure.

Statistical Analysis

Multiple dependent variables were compared. All dependent variables were on the interval scale of measurement. Data were analyzed using independent-samples t test. All t-test comparisons were adjusted to account for disproportionate SDs between groups (equal variances were not assumed). The t-test comparisons were based on 2-tailed probability values, with P<0.05 being considered statistically significant.

This study was approved by the institutional review board at the duPont Hospital for Children.

Results

The BT shunt and RV-PA groups were comparable with regard to gestational age (38.3 versus 38.5 weeks); birth weight (3.2 versus 3.0 kg), age at Norwood operation (4.5 versus 9 days), gender distribution (5/10 versus 9/14 male), presence of aortic atresia (5/10 versus 8/14), and ascending aortic size (2.9±0.9 versus 3.0±1.0 mm), respectively. Age and weight at pre–hemi-Fontan catheterization were also similar for the BT shunt and RV-PA conduit groups (5.2 versus 4.8 months and 5.48 versus 5.45 kg).

Hemodynamic data for the 2 groups are presented in the Table. The patients in the RV-PA conduit group had a higher aortic diastolic pressure (55 versus 42 mm Hg), a narrowed aortic pulse pressure (43 versus 64 mm Hg), a decreased mean pulmonary artery pressure (13 versus 17 mm Hg), a lower Qp:Qs ratio (0.9 versus 1.4), a higher ratio of pulmonary artery to aorta diameter (1.51 versus 1.37), and a higher coronary perfusion pressure (46 versus 32 mm Hg). Aortic oxygen saturation, superior vena cava saturation, RV and aortic systolic pressure, RV diastolic pressure, and pulmonary vascular resistance were not significantly different between groups.

Angiography demonstrated good right ventricular performance in all RV-PA patients without significant tricuspid regurgitation. There were no identified wall motion abnormalities or aneurysm formation at the area of the conduit insertion. With selective pulmonary artery angiography, no appreciable regurgitation was present through the RV-PA conduit into the RV.

Discussion

Over the past two decades, survival for patients undergoing the Norwood procedure for HLHS has increased. Improvements in pre- and postoperative care, refinements in surgical technique, and a greater understanding of the cardiovascular physiology of HLHS have contributed to these results. A commonly used management strategy for these patients has been the limitation of pulmonary blood flow by manipulation of pulmonary or systemic vascular resistance. Techniques have included the addition of carbon dioxide to the inspired gas mixture, use of hypoxic gas, controlled hypoventilation, pharmacological manipulation of systemic vascular resistance, and manipulation of shunt size.

The RV-PA conduit modification created a hemodynamic profile characterized by a higher aortic diastolic pressure, a decreased Qp:Qs, a narrowed systemic pulse pressure, and a higher coronary perfusion pressure. The RV-PA modification was effective in improving coronary blood flow by the limitation of pulmonary blood flow, particularly in diastole.

In our clinical experience after the RV-PA conduit modification, there has been a diminished need for measures to control pulmonary blood flow as well as improved survival.

Patients with the RV-PA conduit are less susceptible to adverse consequences on pulmonary vascular resistance of increased oxygen concentration as a result of the absence of diastolic runoff from the systemic to pulmonary circulation. Ventilation-perfusion mismatch resulting in pulmonary venous desaturation can adversely affect oxygen delivery in the HLHS patient after the Norwood procedure. Limitation of oxygen delivery can be avoided with higher inspired oxygen concentrations in patients with the RV-PA modification.
Reports from the literature describe a minimum coronary perfusion pressure of 20 to 30 mm Hg being necessary for successful resuscitation, whereas normal coronary perfusion pressure in the healthy newborn is 40 to 50 mm Hg. In our study, coronary perfusion pressure in the RV-PA conduit group was 46 mm Hg, whereas it was 32 mm Hg in the BT shunt group, just above the pressure suggested for successful resuscitation. Although there are no data on the importance of coronary perfusion pressure to outcome in patients with HLHS, we believe it reasonable to conclude that the RV-PA conduit modification may have a significant impact on survival between stage I surgery and the hemi-Fontan procedure.

Potential adverse sequelae of the RV-PA modification of the Norwood operation include impaired ventricular systolic or diastolic performance caused by an incision in the right ventricle, diastolic regurgitation through the conduit, and inadequate pulmonary artery growth. These potential complications were not evident, based on similar right ventricle–filling pressures in both groups and no angiographic evidence of wall motion abnormalities or conduit regurgitation. Pulmonary artery growth in the RV-PA group was better and pulmonary vascular resistances were similar in both groups. Potential adverse sequelae of the RV-PA modification of the Norwood operation include impaired ventricular systolic or diastolic performance caused by an incision in the right ventricle, diastolic regurgitation through the conduit, and inadequate pulmonary artery growth. These potential complications were not evident, based on similar right ventricle–filling pressures in both groups and no angiographic evidence of wall motion abnormalities or conduit regurgitation. Pulmonary artery growth in the RV-PA group was better and pulmonary vascular resistances were similar in both groups. Our study did not include patients with 3.5-mm shunts, which provide more restricted pulmonary blood flow than 4.0-mm shunts. Smaller shunts have an increased risk for thrombosis over larger shunts, may not facilitate pulmonary artery growth, and do not overcome the problem of diastolic runoff into the pulmonary arteries. We found that pulsatile flow through the RV-PA conduit facilitated PA growth.

Patients with the RV-PA conduit were considered to be good candidates for the hemi-Fontan procedure, and, in fact, all underwent successful operations.

References

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Circulation. 2003;108:782-784; originally published online August 11, 2003;
doi: 10.1161/01.CIR.0000087338.09589.21

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2003 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

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http://circ.ahajournals.org/content/108/7/782

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